

A case report on Ebstein anomaly

Hepsi Natha

Asst. professor of Medical and Surgical nursing, Teerthanker Mahaveer College of Nursing, Teerthanker Mahaveer University, Moradabad, Uttar Pradesh, India

Abstract

Ebstein anomaly is a congenital malformation of the heart that is characterized by apical displacement of the septal and posterior tricuspid valve leaflets, leading to atrialization of the right ventricle with a variable degree of malformation and displacement of the anterior leaflet. Treatment of Ebstein anomaly is complex and dictated mainly by the severity of the disease itself and the effect of accompanying congenital structural and electrical abnormalities. Treatment options include medical therapy, radiofrequency ablation, and surgical therapy. The case report tells us that Mr. y 22yrs old man with Ebstein anomaly presented with a complaints of exercise is intolerance, Cyanosis, Fatigue and dyspnea since 2 months. On examination widely split first heart sound with loud tricuspid component and soft/absent mitral component in the presence of prolonged PR interval; usually normal second heart sound but may be widely split when pulmonary component delayed due to right bundle-branch block; third and fourth heart sounds commonly present, even in the absence of congestive heart failure. Patient was under the treatment of medical management.

Keywords: Ebstein anomaly

Introduction

Ebstein's anomaly is a rare heart defect that's present at birth (congenital). In Ebstein's anomaly, your tricuspid valve — the valve between the two right hearts chambers (right atrium and right ventricle) — doesn't work properly. The tricuspid valve sits lower than normal in the right ventricle, and the tricuspid valve's leaflets are abnormally formed.

Blood may leak back through the valve, making your heart work less efficiently. Ebstein's anomaly may also lead to enlargement of the heart or heart failure.

Incidence.

The overall prevalence was 0.72 per 10,000 live births. Variables associated with an increased prevalence of non-syndromic Ebstein anomaly included: maternal age >39 years (compared to those 20-24 years), maternal residence along the Texas-Mexico border (compared to non-border residence), and conception in fall or winter (compared to summer). In addition, infants with Ebstein anomaly were at a greater risk of preterm birth and being small for gestational age. These findings help to define subgroups of women at increased risk of having offspring affected by Ebstein anomaly. Furthermore, our findings add to the limited body of literature on this rare but serious malformation.

Symptoms

Mild forms of Ebstein's anomaly may not cause symptoms until later in adulthood. If signs and symptoms are present, they may include:

- Shortness of breath, especially with exertion
- Fatigue
- Heart palpitations or abnormal heart rhythms (arrhythmias)
- A bluish discoloration of the lips and skin caused by low oxygen (cyanosis)

Causes: Ebstein's anomaly is a heart defect that you have at birth (congenital). Why it occurs is still unknown. To understand how Ebstein's anomaly affects your heart, it helps to know a little about how the heart works to supply your body with blood.

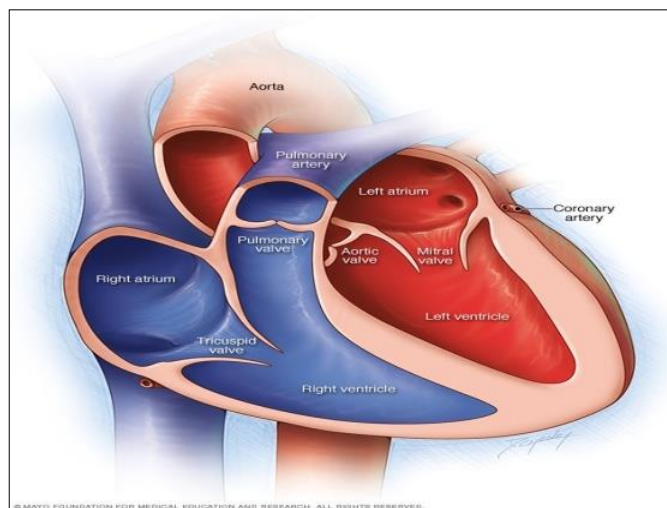


Fig 1

Diagnostic Considerations

Rule out Ebstein anomaly in patients with the following:

- Accessory pathway-mediated WPW syndrome and SVT
- Cyanotic congenital heart diseases
- Atrial septal defect (ASD)
- Cases of severe right heart failure
- Isolated severe tricuspid regurgitation
- L-transposition of the great vessels

Ebstein anomaly should not be confused with Uhl anomaly, a congenital heart disease characterized by a hypoplastic, underdeveloped inflow segment of the right ventricle with tricuspid leaflets arising appropriately from the annulus.

Management treatment of Ebstein's anomaly depends on the severity of the defect and your signs and symptoms. The goal of treatment is to reduce your symptoms and avoid future complications, such as heart failure and arrhythmias. Treatments may include.

Regular monitoring

If you have no signs or symptoms or abnormal heart rhythms, your doctor may recommend only careful monitoring of your heart condition with regular checkups.

Follow-up appointments generally include a physical examination and tests. Tests may include an electrocardiogram, echocardiogram, a Holter monitor test and an exercise stress test.

Medications

If you have heart rhythm disturbances, medications may help control your heart rate and maintain normal heart rhythm.

Your doctor may also prescribe medications for signs and symptoms of heart failure, if needed, such as drugs to prevent water retention (diuretics) and other medications. You also may be given medications to prevent blood clots if you have certain heart rhythm problems or a hole (atrial septal defect) between the upper heart chambers.

Some babies may be given a medication to keep a connection (ductus arteriosus) open between two major blood vessels leading from the heart — the aorta and pulmonary artery. This can help increase blood flow to the lungs. Some babies also may be given an inhaled substance called nitric oxide to help improve blood flow to the lungs.

Surgery

Your doctor may recommend surgery when your symptoms are affecting your quality of life. Surgery may also be recommended if you have mild symptoms but your heart is beginning to enlarge and your overall heart function is beginning to decrease. Because Ebstein's anomaly is rare, choose a surgeon who's familiar with the defect and who has training and experience performing procedures to correct it.

- **Tricuspid valve repair:** In this procedure, surgeons reduce the size of the valve opening and allow the existing valve leaflets to come together to work properly. A band may be placed around the valve to stabilize the repair. This procedure is usually done when there's enough valve tissue to allow for repair.

Self-management

By Mayo Clinic Staff Print

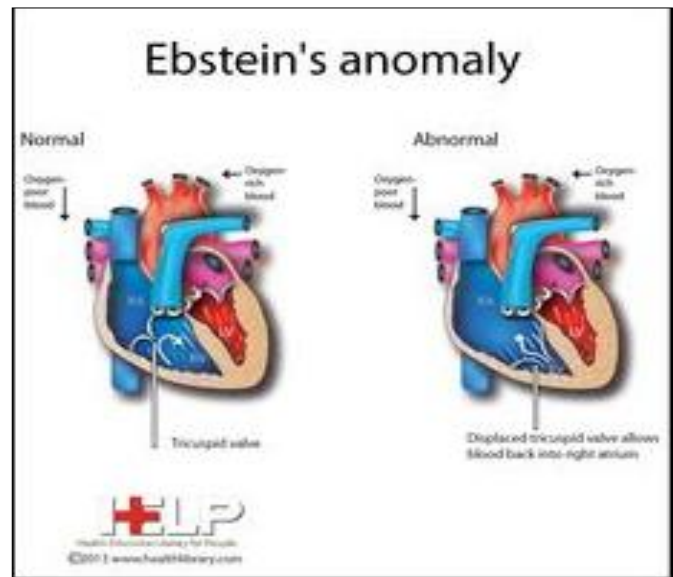
Coping and support

If you or your child has mild Ebstein's anomaly, here are some steps that may help you cope:

- **Follow up on medical care:** Be sure to follow up with your cardiologist trained in congenital heart disease for regular evaluations. Be an active participant in monitoring the condition and report any new or worsening signs or symptoms to your doctor. Timely treatment can keep the condition from becoming worse.

- **Take medications as prescribed:** Taking medications at the right dose and the right time can help improve symptoms such as racing heartbeats, fatigue and shortness of breath.
- **Stay active:** Be as physically active as your or your child's doctor allows. Exercise can help strengthen the heart and improve blood circulation. If you're a parent of a child with Ebstein's anomaly, it's natural to want to protect your child from harm. But remember that your child needs to live life as normally as possible. Encourage playtime with breaks as needed. Ask your doctor for a note you can give to your child's teachers or caregivers describing any restrictions on his or her physical activity.
- **Develop a support network:** Although many people with congenital heart defects lead normal, healthy lives, living with a heart defect isn't always easy, particularly when you or your child needs continued specialized care. The physical, emotional and financial stress of coping with a serious health condition can be overwhelming.

Having family and friends you can rely on is critical to successful coping. In addition, you may wish to ask your doctor about local support groups that may be helpful. Support groups can be a great source of practical information, comfort and friendship.



Case Report

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- Antibiotic prophylaxis for infective endocarditis
- Medical therapy for heart failure - Angiotensin-converting enzyme (ACE) inhibitors, diuretics, and digoxin

Reference

1. Muñoz-Castellanos L, Espinola-Zavaleta N, Kuri-Nivón M, Keirns C. Ebstein's Anomaly: anatomico-echocardiographic correlation. *Cardiovasc Ultrasound*. 2007; 23:5-43.
2. Armengol Rofes AJ, Serrano Durán M, Albert Brotons DC, Sánchez López C, Casaldáliga Ferrer J, Girona Comas JM. [Ebstein's anomaly of the tricuspid valve. Apropos 35 cases. *An Esp Pediatr*. 1996; 44(2):139-44.
3. Khositseth A, Khowsathit P. Factors affecting mortality in Ebstein's anomaly of the tricuspid valve. *J Med Assoc Thai*. 1999; 82(1):S10-5.
4. Correa-Villaseñor A, Ferencz C, Neill CA, Wilson PD, Boughman JA. Ebstein's malformation of the tricuspid valve: genetic and environmental factors. The Baltimore-Washington Infant Study Group. *Teratology*. 1994; 50(2):137-47.
5. Boston US, Goldberg SP, Ward KE, *et al*. Complete repair of Ebstein anomaly in neonates and young infants: A 16-year follow-up. *J Thorac Cardiovasc Surg*. 2011; 141(5):1163-9.
6. Attenhofer Jost CH, Connolly HM, Scott CG, Burkhart HM, Warnes CA, Dearani JA. Outcome of cardiac surgery in patients 50 years of age or older with Ebstein anomaly: survival and functional improvement. *J Am Coll Cardiol*. 2012; 59(23):2101-6.
7. Attie F, Casanova JM, Zabal C, Buendía A, Miranda I, Rijlaarsdam M. Ebstein's anomaly. Clinical profile in 174 patients. *Arch Inst Cardiol Mex*. 1999, 69(1):17-25.
8. Brown ML, Dearani JA, Danielson GK, Cetta F, Connolly HM, Warnes CA, *et al*. Effect of operation for Ebstein anomaly on left ventricular function. *Am J Cardiol*. 2008 102(12):1724-7.
9. Brown ML, Dearani JA, Danielson GK, Cetta F, Connolly HM, Warnes CA, *et al*. Functional status after operation for Ebstein anomaly: the Mayo Clinic experience. *J Am Coll Cardiol*. 2008; 52(6):460-6.